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Karolinska Institutet, Stockholm, Sweden

# **CHILDREN WITH LOW ANORECTAL MALFORMATIONS AND THEIR PARENTS: QUALITY OF LIFE, PHYSICAL AND PSYCHOSOCIAL FUNCTION**

Helena Wigander



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**Children with low anorectal malformations and their parents:  
Quality of life, physical and psychosocial function**

## **AKADEMISK AVHANDLING**

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Av

**Helena Wigander**

*Huvudhandledare:*

Med dr Maria Öjmyr-Joelsson  
Karolinska Institutet  
Institutionen för Kvinnor och Barns Hälsa

*Bihandledare*

Med dr Margret Nisell  
Röda Korsets Högskola  
Hälsovetenskapliga Institutionen

Professor Björn Frenckner  
Karolinska Institutet  
Institutionen för Kvinnor och Barns Hälsa

Professor Tomas Wester  
Karolinska Institutet  
Institutionen för Kvinnor och Barns Hälsa

*Fakultetsopponent:*

Professor Mats Eriksson  
Örebro Universitet  
Institutionen för Hälsovetenskaper

*Betygsnämnd:*

Docent Gundela Holmdahl  
Göteborgs Universitet  
Institutionen för Kliniska Vetenskaper

Docent Björn Wrangsjö  
Karolinska Institutet  
Institutionen för Kvinnor och Barns Hälsa

Docent Kristina Burström  
Karolinska Institutet  
Institutionen för Lärande, Informatik, Management  
och Etik

Till mina söner

*Livet måste levas framifrån men kan endast förstås bakifrån*

-Kierkegaard

## ABSTRACT

Anorectal malformations (ARM) are congenital anomalies affecting 1 in 3 000 births. The malformation varies from very minor to complex and can be divided in to low, intermediate, and high form of ARM. Children with a low ARM are primarily repaired in the newborn period. The children often have some degree of functional problems such as constipation and incontinence. Depending on the severity, laxatives and sometimes bowel irrigations are necessary to obtain and maintain an acceptable bowel function. Impaired bowel function might also lead to impaired Quality of Life (QoL) and poor psychosocial functioning.

The aim of this thesis was to assess QoL, bowel function and psychosocial function in children and adolescents with low ARM. The aim was also to investigate parenting stress in parents of children and adolescents with low ARM. An additional aim was to translate and culturally adapt the disease-specific QoL questionnaire Hirschsprung's Disease/Anorectal malformation Quality of Life Questionnaire (HAQL) into Swedish.

Forty-four children with low ARM born between 1993 and 2007 who had been operated on, were asked to participate along with their parents.

Study I presented a methodological study design using a forward- back translation technique and cultural adaption of the translated HAQL. A pilot study with patients with Hirschsprung disease was also performed. The English version was not accurately translated, but the Swedish translation was found to correlate well with the original Dutch version.

In Study II, QoL and bowel function were assessed in children and adolescents with low ARM using a descriptive comparative design with a retrospective data collection using two questionnaires the: HAQL and Bowel Function Score (BFS). Seventeen patients and 18 mothers completed the HAQL. Twenty-seven families completed the BFS. Healthy controls with 17 children also completed the HAQL. The children showed impaired function in three HAQL domains when compared to controls. Compared with their mothers, they reported impaired physical function and more symptoms in the emotional functioning and physical symptom domains. Of the families who completed the BFS, 63% reported normal bowel function, 33% moderate function and 4% reported poor function. An evaluation of the HAQL of the Fecal Continence (FC), Emotional Functioning (EMF) and Physical Function (PH) domains showed no obvious conflicts.

Study III assessed psychosocial function in children and adolescents with low ARM using a descriptive comparative design using two questionnaires the Imperforate Anus Psychosocial

Questionnaire (IAPSQ) and the Child Behavior Check List (CBCL). Sixteen children from the index group described completed the IAPSQ and 18 mothers from the index group completed the CBCL. A control group with 17 healthy children as well as an additional control group consisting of 25 children with intermediate and high ARMs completed the IAPSQ. Seventeen mothers from the control group of healthy children also completed the CBCL. The children with low ARM seemed to have good psychosocial function. When compared to the control group of children with high- and intermediate ARM, differences were found in one domain, Emotional/Cognition (COG), where the control group had lower function. The majority of the children and adolescents scored by their mothers in the CBCL were within normal range of the syndrome scale. In total, five children (33%) were scored within borderline or clinical range in one or several of the problem scores on the syndrome scale.

Study IV investigated parenting stress in parents of children and adolescents with low ARM using a sequential explanatory mixed-method design, including both quantitative and qualitative methods with questionnaires and interviews. Fifteen mothers and 13 fathers completed the Swedish Parenthood Stress Questionnaire (SPSQ) and, semi-structured interviews were performed with 9 parents from the index group. A control group consisting of the parents (17 mothers and 6 fathers) of healthy controls was used for comparison in the quantitative strand. There were no differences in stress between the index group and control group except for in one subscale; INCOMPETENCE, where the index group reported lower stress levels compared to controls. The interviews revealed three themes: *Communication between parents*, *Expectations of parenthood*, and *Challenges concerning parenthood*, where the parents told about early experiences of emotional stress with feelings of guilt and chaos at the time of birth and during infancy.

# POPULÄRVETENSKAPLIG SAMMANFATTNING

Analatresi är en medfödd missbildning som drabbar 1 av 3000 födda barn. Missbildningen varierar från lindrig till svår och kan delas in i låg, intermediär och hög form. Barn med låg analatresi opereras under nyföddhetsperioden. Även om barnen opereras har de ofta problem med förstoppning och läckage. Beroende på missbildningens svårighetsgrad är behandling med mjukgörande läkemedel och ibland behandling med lavemang nödvändigt för att uppnå och bibehålla en acceptabel tarmfunktion. Att ha en nedsatt tarmfunktion kan leda till en försämrad livskvalitet och en dålig psykosocial funktion.

Syftet med avhandlingen var att undersöka barnens livskvalitet, tarmfunktion och psykosociala funktion samt att undersöka föräldrastress hos föräldrarna. Vi ville också översätta och kulturellt anpassa det sjukdomsspecifika frågeformuläret HAQL till svenska förhållanden.

I delstudie II-IV har 44 barn och ungdomar med låg analatresi som var födda mellan 1993 och 2007 och som hade opererats och vårdats på Astrid Lindgrens barnsjukhus och deras föräldrar tillfrågats om att delta i studien.

Studie I är en metodstudie med en översättning av frågeformuläret HAQL. En översättningsmetod med en så kallad ”*forward back translation technique*” vilket innebär att man översätter till målspråket och sen översätter tillbaka till ursprungsspråket. Även en kulturell anpassning av det översatta frågeformuläret gjordes. Vidare utfördes en pilotstudie med patienter med sjukdomen Hirschsprung. Översättningen av den engelska versionen visade sig inte vara korrekt utförd. Detta ledde till att ytterligare en översättning till originalspråket holländska utfördes, den senare översättning visade att den svenska versionen överstämde väl med den holländska originalversionen.

I studie II utvärderades tarmfunktion och livskvalitet hos barnen och ungdomarna. Två formulär användes i denna studie: Det formulär som hade översatts i studie I, HAQL samt ett formulär som mäter tarmfunktion BFS. Av de tillfrågade familjerna valde 17 barn/ungdomar och 18 mödrar att fylla i formulären. Tjugosju familjer fyllde även i BFS. Jämfört med friska kontroll patienter hade barnen/ungdomarna en sämre funktion i tre av HAQLs domäner. Jämfört med sina mammor skattade barnen/ungdomarna en sämre fysisk funktion och mer emotionella besvär. Av familjerna som fyllde i BFS rapporterade 63% en normal tarmfunktion, 33% uppgav måttliga besvär och 4 % hade dålig tarmfunktion. En utvärdering

av domänerna Fecal Continence (FC), Emotional Functioning (EMF) och Physical Function (PH) visade inte på några uppenbara motsättningar, och de utvärderade domänerna verkar mäta det som de är avsedda att mäta.

I Studie III undersöktes psykosocial funktion hos barnen/ungdomarna. Två formulär användes formuläret IAPSQ som är ett sjukdomsspecifikt frågeformulär som mäter psykosocial hälsa samt formuläret CBCL som skattar beteende och emotionell funktion hos barn och ungdomar. Sexton barn och ungdomar från indexgruppen fyllde i IAPSQ och 18 mödrar fyllde i CBCL. Sjutton friska barn och ungdomar fyllde i IAPSQ som kontroll. Ytterligare en kontrollgrupp med 25 barn med en svårare form av analatresi som fyllt i IAPSQ vid ett tidigare tillfälle användes som jämförelse. Även 17 mödrar till barn/ungdomar i den friska kontrollgruppen fyllde i CBCL som jämförelse. Resultatet visade att barnen/ungdomarna med låg analatresi hade en bra psykosocial funktion. Jämfört med kontrollgruppen med den svårare formen av analatresi visade det sig att kontrollgruppen skattade en sämre psykosocial funktion gällande domänen Emotional/Cognition (COG). Majoriteten av barnen/ungdomarna som skattades av sina mammor med instrumentet CBCL befanns vara inom det normala på syndromskalan. Totalt skattades 5 barn (33%) vara inom borderline eller klinisk i flera av problem-skalorna på syndromskalan.

I den fjärde studien undersöktes föräldrastress hos föräldrar till barn med låg analatresi. Både enkäter och intervjuer användes i med en så kallad "mixed-method design". Femton mammor och 13 pappor fyllde i ett formulär som skattar föräldrastress. The Swedish Parenthood Stress Questionnaire (SPSQ). Även intervjuer med nio föräldrar genomfördes. Som jämförelse fyllde även 17 mödrar och 6 fäder till friska barn i SPSQ. Resultatet visade att föräldrarna till barnen med låg analatresi skattade låga nivåer av stress, det var inga skillnader i stressnivåer mellan föräldrarna och kontrollgruppen förutom skalan INCOMPETENCE där indexgruppen rapporterade en lägre stress än kontrollgruppen. Intervjuerna avslöjade tre teman: *Kommunikation mellan föräldrar, Förväntningar på föräldraskapet och Utmaningar i föräldraskapet*. Föräldrarna berättade om tidiga erfarenheter av emotionell stress med känslor av skuld och kaos som uppkom i samband med barnets födelse och under spädbarnstiden.



# FÖRORD

År 2005 började jag arbeta på tarmterapimottagningen som specialistsjuksköterska för barnen som är födda med analatresi. Jag fick då förmånen att börja följa barnen och deras föräldrar och familjer från födelse tills dess de lämnar barnsjukhuset i tonåren, detta efter att tidigare endast träffat familjerna i slutenvården. Detta var så mycket mer, det gav en helt annan förståelse för att leva med en kronisk sjukdom, men väckte också en nyfikenhet för att få veta mer. Tarmterapimottagningen var och är en dynamisk plats där forskning rörande denna patientgrupp stod i fokus och som jag snart blev inbjuden till att delta i. Det dröjde dock ytterligare några år innan jag påbörjade det forskningsprojekt och den resa som nu sammanfattas i denna avhandling.

Detta arbete hade aldrig kunnat genomföras utan engagerade föräldrar och barn samt ungdomar som valt att dela med sig av sina erfarenheter, sin kunskap och sina funderingar. Detta i förhoppning om att vi tillsammans skulle kunna öka kunskapen kring hur det är att leva med en medfödd missbildning och att detta ska komma andra familjer till gagn.

Astrid Lindgrens Barnsjukhus, 10 oktober, 2018

Helena Wigander

## LIST OF SCIENTIFIC PAPERS

This thesis is based on the following four papers, which will be referred to in the text by their Roman numerals (I-IV).

- I. **Wigander H**, Frenckner B, Wester T, Nisell M, Öjmyr-Joelsson M.  
Translation and cultural adaption of the Hirschsprung's Disease/Anorectal Malformation Quality of life Questionnaire (HAQL) into Swedish.  
*Pediatric Surgery International*, 2014, Vol.30(4), pp.401-406
- II. **Wigander H**, Nisell M, Frenckner B, Wester T, Brodin U, Öjmyr-Joelsson M.  
*Quality of life and functional outcome in Swedish children with Low Anorectal Malformations. A follow-up study.*  
Manuscript submitted
- III. **Wigander H**, Nisell M, Wester T, Frenckner B, Öjmyr-Joelsson M.  
*Psychosocial function in children and adolescents with low Anorectal malformations.*  
In manuscript
- IV. **Wigander H**, Öjmyr-Joelsson M, Frenckner B, Wester T, Nisell M.  
Impact of Low Anorectal Malformation on Parenting Stress: A Mixed-Method Study.  
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## LIST OF ABBREVIATIONS

ARM	Anorectal malformation
BFS	Bowel Function Score
CBCL	Child Behavior Check List
HAQL	Hirschsprung/Anorectal malformation Quality of Life Questionnaire
HD	Hirschsprung Disease
HRQoL	Health Related Quality of Life
IAPSQ	Imperforate Anus Psychosocial Questionnaire
SD	Standard deviation
SPSQ	Swedish Parenthood Stress Questionnaire
QoL	Quality of Life

# 1 INTRODUCTION

## 1.1 ANORECTAL MALFORMATIONS

Anorectal malformations (ARMs) are congenital anomalies involving the anus and rectum. The incidence of ARM is 1 in 3 000 live births and affects both boys and girls but is slightly more frequent among boys [1-3]. ARMs represent a wide spectrum of anomalies ranging from mild to more severe and vary considerably in complexity and functional prognosis.

The Wingspread International Classification originates from the Wingspread conference in 1984 and distinguishes high, intermediate and low malformations, with special groups for cloacas' and rare malformations. The Wingspread classification was a commonly used classification system until 2005 when the Krickenbeck conference was held. The most recent classification system is the Krickenbeck International Classification system of ARM, which is a consensus recommendation between leading international authorities and is based upon the Wingspread classification as well as Pena's classification. The goal of the conference was to develop standards for a classification of ARM based on the fistula location, cloacal lesions, lesions with no fistula, anal stenosis and other rare variants. The Krickenbeck Classification (Table 1) enables the different operative procedures to be more comparable. The Wingspread Classification is considered useful when choosing surgical approach [4, 5]. In this thesis low ARM refer to perineal fistula and imperforate anus without fistula [6]

Associated anomalies are encountered in more than 60% of the patients and include renal, cardiac, gastrointestinal, vertebral, skeletal, genitourinary and gynecologic anomalies, developmental delay and chromosomal defects [5, 7, 8].

Lower urinary tract dysfunction with incontinence is common in children with more severe types of ARM [9]. However, children and adolescents with low ARM did not differ from control patients in prevalence of Lower Urinary Tract Symptoms (LUTS) [10, 11].

### 1.1.1 Surgical treatments

During the first 24 hours after birth a thorough investigation is done to exclude other associated malformations and to gather information in order to decide whether to do a primary repair or to perform a colostomy. A colostomy is usually performed in newborn babies with a high ARM. The colostomy provides decompression of the bowel and is useful

to further investigate the malformation with a colostogram [1]. Posterior sagittal anorectoplasty (PSARP) was first described by de Vries and Pena in 1980 and is the operative procedure of choice. The baby is placed in a prone position and the sphincter is located by electric stimulation. The incision is made in the midline and the sphincter is divided. The rectum is identified, dissected, and opened and the fistula is localized. The rectum is then mobilized to reach the center of the sphincter complex [1, 5, 12]. A laparoscopy approach combined with the PSARP can be used when the rectum is very high and difficult to reach without laparotomy [13, 14]. Limited PSARP is used in cases of perineal fistula and consists of a small posterior sagittal incision with enough mobilization of the fistula to be transposed and placed within the sphincter [1].

In order to achieve a satisfactory function, the anus needs to be dilated. Two weeks after surgery, dilatations of the anus are initiated. The dilatations are performed by the parents according to an anal dilatation program twice a day until the desired size is reached [8].

### **1.1.2 Bowel function**

Children with ARM lack a normal anal canal, have varying degrees of sphincter insufficiency, and have motility disorders in the colon [15]. Further, associated malformations such as sacral defects and spinal abnormalities also have an influence on bowel function [8]. As a result, ARM patients often have functional problems such as constipation or incontinence [16]. A study on adolescents with ARM where all sub-groups were included showed considerable intestinal symptoms such as soiling, constipation and gas incontinence in all subgroups [17].

Because constipation and fecal incontinence are common problems, many patients depend on regular treatments, such as enemas and oral laxatives. The treatment is usually required for several years [1]. To be able to achieve an optimal outcome, it is important to detect and treat complications such as constipation early [7]. Poor bowel function was found to be common in early childhood, but the functional outcome concerning continence, soiling, and constipation improved with age [18, 19]. It is important to have regular follow-ups to ensure compliance with treatment and to prevent secondary complications [20].

Long-term results have been considered to be good in the majority of the patients with low ARM. However, it is common for children with low ARM to have problems with

constipation [6, 21]. Long-term bowel function has been shown to be impaired in one-third of the patients with a low ARM, mainly constipation and/ or soiling. In most of the patients, the constipation and the soiling are mild and do not cause any social problems [6].

Gender differences in outcome have been shown, and males born with perineal fistulas tend to have less incontinence and constipation than females born with perineal fistulas [22]. Studies conducted in Finland on low ARM patients showed similar results in gender differences, and female patients born with anterior anus and male patients born with low ARMs were comparable to healthy controls in adulthood. Two thirds of female patients born with perineal or vestibular fistulas are likely to achieve bowel function equivalent to healthy peers and the majority of male patients with perineal fistulas will achieve socially acceptable continence [10, 11, 23].

About 75% of ARM patients have the potential for continence when using medical strategies to prevent and treat constipation. The remaining 25% might benefit from daily washouts and antegrade enemas to manage fecal incontinence [24]. Compliance with bowel management is the key to being able to achieve better bowel function [1]. A study in patients with high and intermediate ARM showed that it was possible to achieve social continence by age 5 with individualized support [18].

For patients with fecal incontinence a bowel management program with daily enemas is a way to keep themselves clean [25]. The program is individualized for each patient and is described as an” ongoing” process of trial and error that responds to the individual patient”. The program is carried out for a week with daily abdominal radiographs to be able to tailor the regime. It has been reported that more than 95 % of the children are clean and dry when following this program. [15].

Antegrade enema through an appendicostomy was first described in 1990 as an alternative to rectal irrigations. The appendix is used to create a continent stoma [26]. The use of antegrade enema may increase the child’s autonomy [27], and good outcomes have been reported regarding fecal continence and improvement in quality of life (QoL) [28, 29].

There are several different scoring methods to evaluate bowel functions such as constipation and fecal continence in ARM patients. For example, The Krickenbeck criteria and the Bowel function score (BFS) [4, 30] are both validated and commonly used instruments.



## **1.2 QUALITY OF LIFE**

QoL can be described as a holistic and broad concept and concerns an individual's satisfaction in all different aspects of life and is a multidimensional assessment of a person's current life situation [31, 32]. QoL has been defined by the WHO as a "broad multidimensional concept which includes subjective evaluations of both positive and negative aspects of life [33].

### **1.2.1 Health related quality of life (HRQoL)**

When assessing a person's health status, the concept of health-related quality of life (HRQoL) is used. HRQoL focuses on health care aspects of a person's life [34] and is defined as a multidimensional concept that covers social, physical, and psychological domains of health [32, 35, 36]. HRQoL is an important concept and helps us to improve our understanding of children with disabilities. Assessment of children's HRQoL is necessary to be able to obtain a complete health status [31, 37].

To assess a person's HRQoL, patient-reported outcome measures are used. HRQoL is subjective, and when possible it, should be assessed from the patient's perspective [38]. It is well established that pediatric self-reports should be used as the standard when measuring HRQoL and other symptom measurements used in children [39] because patient self-reports reported by children are not synonymous with proxy-reported information [40]. Studies have shown that proxies tend to underestimate patients' HRQoL, and it is common for observers to underestimate the impact of psychological aspects and to focus more on physical signs [41-43].

Even though self-reports should be considered standard when measuring HRQoL in children, there are situations where a child might be too young or too ill to be able to complete a HRQoL instrument. In such cases parent-proxy reports may be needed [40]. The parent proxy-report should also be considered as a secondary measure because of parents' roles concerning clinical decision-making for chronically ill children [44]. By using both proxy- and children's self-reports when assessing children's HRQoL, valuable complementary information might also be provided [37, 45].

When choosing an HRQoL instrument there are two types of instrument to consider – generic instruments and disease-specific instruments. Generic instruments are intended for a more

general use. The generic instrument covers a wide range of conditions and a general population and has the advantages that it enables comparisons across pediatric populations [32, 40]. Examples of generic pediatric self-assessment instruments available in Swedish are the Pediatric Quality of Life Inventory (PedsQoL) and KIDSCREEN [46, 47]

To be able to assess HRQoL in a disease-specific population, a self-reported measurement is required that is consisting of items that is related to physical, emotional and social functioning, as well as disease-related symptoms [48]. Disease-specific instruments are considered sensitive and focus on issues that concern specific problems. The disease-specific instrument may enhance measurement sensitivity for health domains closely linked to a specific chronic health condition [40]. Because there were no disease-specific instruments available in Swedish for children and adolescents with ARM, we decided to translate and culturally adapt the disease-specific quality of life questionnaire, the Hirschsprung's Disease/Anorectal Malformation Quality of Life Questionnaire (HAQL) [48], into Swedish.

### **1.2.2 Translation and cultural adaption of a HRQoL instrument**

The translation of a HRQoL instrument using a forward-back translation technique is a process including several steps such as translation into the target language by two independent translators. A committee works together with cultural adaption of the translation, a back translation to the original language and a comparison of the original version and the final translation. There are several different translation methods and guidelines presented in the literature [49-53]. However, no consensus on one translation method has yet been established.

To achieve a cross-culturally comparable version of a HRQoL instrument, a cross-cultural adaptation of the instrument is necessary. Cross-cultural adaption describes the process of preparing an instrument for use in a different setting [54]. Cross-cultural adaption involves both linguistic translation where the instrument is literally translated into the new language, and cultural adaption, where it is adjusted to make it appropriate for the cultural context [55].

### **1.2.3 Quality of life in ARM patients**

The concept of HRQoL in children and adolescents with ARM has been described in several studies using different instruments, both generic and disease-specific with various reported outcomes. When using generic questionnaires, no differences were found in reported HRQoL between children with ARM and healthy controls [56]. However, others reported differences with lower experienced HRQoL in both children and adolescents compared to healthy children [57, 58]. When using the disease-specific QoL questionnaire HAQL on children and adolescents with ARM, low QoL levels were found in the emotional and physical functioning domains [59].

Patients with ARM reported slightly lower QoL than comparison groups, and when compared to adolescents, children report better QoL but worse disease-specific functioning [60]. Even though the bowel function in most ARM patients is not considered to be the same as in healthy peers, most patients report a good QoL without social restrictions [7]. In contrast, the patients' bowel function was found to impact QoL in ARM patients when compared to healthy controls [61]. Furthermore, in adult patients with ARM psychosocial functioning was found to be the factor that had the greatest impact on QoL [62].

## **1.3 PSYCHOSOCIAL FUNCTION**

Psychosocial function concerns many different aspects and can be defined as behavioral, emotional, and educational issues that are highly prevalent among children and can interfere with everyday functioning [63]. The physical consequences, such as fecal incontinence, in children born with ARM might cause psychological problems. The part of the body that is involved and the characteristics of the malformation are especially relevant. The child has to participate and allow treatment that means that the child's integrity is not fully respected and might influence the relationship between the child and the parents [64, 65].

Previous studies of children with high and intermediate ARM at Astrid Lindgren Children's Hospital showed above all that these children had a good psychosocial function. Functional bowel problems did not make the children feel limited in school. Parents to children with imperforate anus had an important role and were very involved in the follow up treatment [66, 67].

The Imperforate Anus Psychosocial Questionnaire (IAPSQ), a self-report questionnaire, has been validated in children with ARM and is designed to measure the children's and adolescents' psychosocial function. The questionnaire is also available in a proxy version [68]. The Child Behavior Checklist (CBCL/6-18) measures children's and adolescents' emotional and behavioral problems and is scored by the children's/adolescents' parents.

## **1.4 PARENTING STRESS**

Parenting stress is defined as stress that, is directly related to the role of being a parent and can have a variety of different effects on the parents and their children [69]. Parenting stress can affect several different areas of life and may have negative effects on the relation between the child and the parent. This can lead to negative consequences for the child's development and behavior. Factors contributing to higher levels of stress are social background, employment, levels of education, and being a mother, and these are also contributing factors for higher levels of parenting stress [70]. Parenting stress can also affect child health-related outcomes and might interfere with the management of a child's chronic condition [71].

Parents of chronically sick children reported higher stress levels than parents of healthy children [72, 73], and mothers, especially single mothers of children with rare diseases scored high for parental stress and physical and emotional strain during the child's first year [74].

There appear to be few studies investigating parental stress and parent's QoL in children with ARM. One study showed that mothers of newborns with ARM and Hirschsprung's disease suffered from more anxiety and a lower psychological QoL than fathers, but the anxiety among mother's decreased when the child became older [75]. Fecal incontinence may affect parent's QoL negatively [76]. Parents of older incontinent children with ARM experienced relatively more stress especially in parents of male children [72].

It is important to be aware of the specific problems related to having a child with a chronic condition. Parents' satisfaction increases with specific information concerning the condition, and health care professionals play an important role in supporting the parents' well-being. The parents have a large responsibility and are very much involved in the treatment of the child both in performing anal dilatations and in preventing and treating constipation and fecal leakage during the child's upbringing [67, 72]. The parents described difficult experiences (such as stress) associated with the malformation, which might have affected all family

members [77]. Support to parents of children with ARM should be individualized for each family, and if necessary experts from child and adolescent psychiatry should be brought in [78].

The Swedish Parenthood Stress Questionnaire (SPSQ) measures parents' perception of stress in their role as parents. The SPSQ is based on the American Parenting Stress Index [79]. The SPSQ is translated and adjusted for Swedish settings [80].

# THESIS AT A GLANCE

	Study Aims	Method/design	Results	Conclusion
I	To translate and culturally adapt the disease-specific instrument, HAQL into Swedish	A methodological study design with a forward back translation technique and cultural adaption of the translated questionnaire. A pilot study with Hirschsprung disease patients and their parents was also performed.	The English version was found to be flawed. The translation correlated well with the original Dutch version. The Swedish translators chose to use a simplified language for the children but not for the adolescent- and proxy versions.	The translation of the HAQL into Swedish provides a disease-specific QoL instrument for children and adolescents with ARM and HD.
II	To investigate the quality of life and bowel function and to evaluate the HAQL questionnaire.	Descriptive comparative design with a retrospective data collection using questionnaires. 17 patients and 18 mothers completed the HAQL. 27 families completed the BFS. Healthy controls with 17 children also completed the HAQL. An evaluation of the HAQL questionnaire using Mokken- and Rasch analysis was also performed, as well as a drop-out analysis.	<p>The children had impaired function in three domains when compared to controls. Compared with their mothers, they reported impaired physical function and more symptoms in the emotional functioning and physical symptom domains.</p> <p>Of the families who completed the BFS 63% reported normal bowel function, 33% reported moderate function and 4% , reported poor function. The evaluation of the HAQL of the FC, EMF and PH domains showed no obvious conflicts.</p>	The children did not differ much regarding their QoL, although they seemed to have impaired bowel function and worse emotional functioning compared to controls. The mothers underestimated their children's physical symptoms and overestimated their emotional functioning. Evaluated domains in the HAQL seem to work as intended. However, the questionnaire needs further development
III	To examine psychosocial functioning and possible emotional and behavioral problems.	Descriptive comparative design with retrospective data collection using the questionnaires IAPSQ and CBCL. 16 children from the index group completed the IAPSQ and 18 mothers from the index group completed the CBCL. The same control group as in study II with 17 children as well as an additional control group consisting of 25 children with intermediate and high ARMs completed the IAPSQ. 17 mothers from the control group also completed the CBCL.	The children scored high overall in the psychosocial domains in the IAPSQ. When compared to the control group of children with high- and intermediate ARM, differences were found in one domain COG, where the control group had lower function. The majority of the children and adolescents scored by their mothers in the CBCL were within the normal range of the syndrome scale. In total, five children, (33%) were scored within the borderline or clinical range in one or several of the problem scores on the syndrome scale	Overall, the children appear to have good psychosocial function, even if some of the children might have some emotional and behavioral problems. It appears that the severity of the malformation might be related to psychosocial function.
IV	To investigate parenting stress among the parents.	Sequential explanatory mixed-method design using both quantitative and qualitative data: 15 mothers and 13 fathers completed the SPSQ and semi-structured interviews with 9 parents from the index group were conducted. A control group consisting of the parents (17 mothers and 6 fathers) of healthy controls from study II and III was used for comparison in the quantitative strand.	There were no significant differences between the index group and control group except for in the subscale of Incompetence, where the index group reported lower stress levels compared to controls. The interviews revealed three themes: <i>Communication between parents</i> , <i>Expectations of parenthood</i> , and <i>Challenges concerning parenthood</i> .	The index group parents reported low levels of stress. When interviewed they told about early experience of emotional stress with feelings of guilt and chaos at the time of birth and during infancy.



## 2 AIMS OF THE THESIS

The overall aim was to investigate how children with low ARM and their parents are affected by the malformation.

The specific aims were to:

- To translate and culturally adapt a disease-specific instrument, the HAQL (Hirschsprung's Disease/Anorectal Malformation Quality of Life Questionnaire), into Swedish (*Study I*).
- To investigate the QoL and bowel function and to evaluate the HAQL questionnaire (*Study II*).
- To examine psychosocial functioning and possible emotional and behavioral problems (*Study III*).
- To investigate parenting stress among the parents (*Study IV*).

The following research questions in particular were addressed:

How can a disease-specific QoL questionnaire be translated and adapted for children with ARM, living in Sweden? (*Study I*). How is the bowel function of children with low ARM? How is the QoL of children with low ARM? (*Study II*). Do the children have behavioral/emotional problems? (*Study III*). What impact does the severity of the malformation have on the child's physical and psychosocial function? (*Study III*) Are there discrepancies between child self- report and parental report on the child's physical and psychosocial functioning? (*Studies II and III*). To what extent do parents take responsibility for the follow-up treatment? How do parents of children with low ARM describe their stress and how do they cope with difficulties related to the malformation? (*Study IV*)





## 3 PATIENTS AND METHODS

### 3.1 PARTICIPANTS

#### 3.1.1 Study I

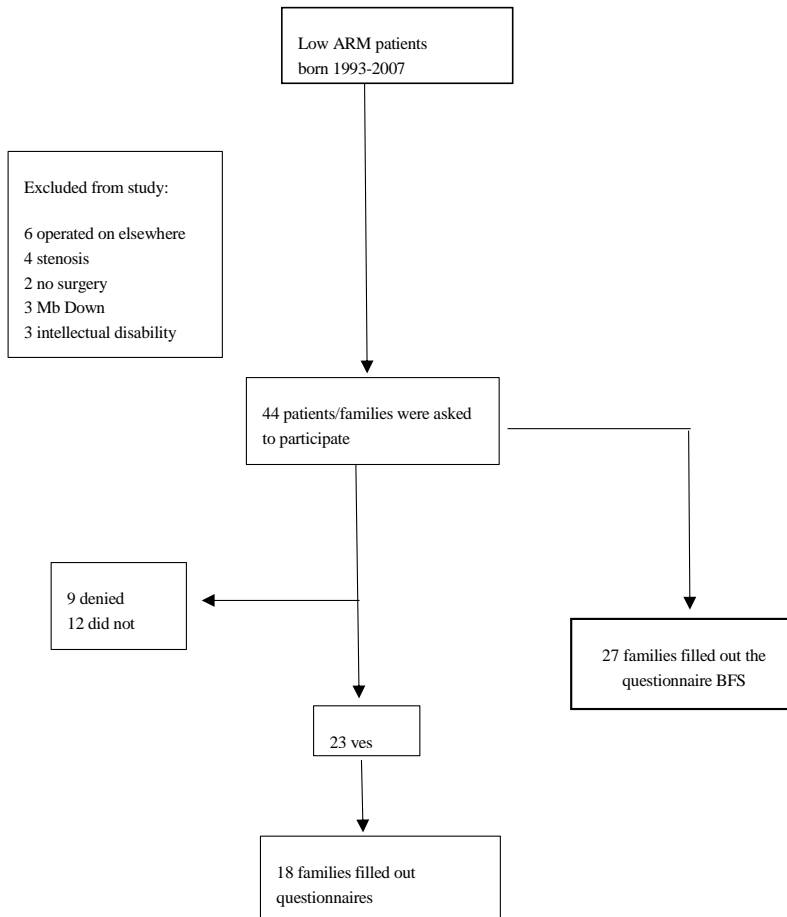
The first paper included patients born with Hirschsprung's disease. Nine children and adolescents, three from each of the age groups: 8 to 11, 12 to 16, and 17 to young adults, were randomly selected from all children with Hirschsprung disease (HD) treated at Astrid Lindgren Children's Hospital, to participate in the pilot study. The families were contacted by postal mail and asked to participate. Parents of the participants from the two younger age groups were asked to complete the proxy version. Six families agreed to participate, two in each age group. One family declined and two of the families did not respond.

#### 3.1.2 Studies II – IV

For study II, III and IV, all children and adolescents with low ARMs born between 1993 and 2007, who had been treated at Sankt Görans and Astrid Lindgren Children's Hospital in Stockholm, Sweden, and their parents were considered for participation. Sixty-four patients were identified. We included patients who had been operated on at our hospitals and were able to complete the questionnaires. In total, 44 families were considered for participation. Twenty-three (53%) families agreed, nine (20%) declined, and twelve families (27%) did not reply. Data were collected during the period 2011 to 2015 and the data collection was extended twice during this period. The children were between the age of 8 and 18 when they were invited to participate. Please see flowchart of participants (figure 1). Table 1 presents clinical data of index group.

**Table 1** Clinical data index group

Associated malformations:	All patients Index group n=44	Participants study II, III, IV n=18	Participants BFS study II n=27	Non-responding (denied or did not answer) n=16
Esophagus atresia	5	2	3	3
Cardiac	3	0	0	3
Renal malformations	2	2	2	0
Lumbar vertebrae	2	1	2	0
Limb	1	1	1	0



**Figure 1.** Flowchart over participants in index group

Fifty healthy children and adolescents who had visited Astrid Lindgren Children's hospital for a minor procedure not affecting the bowel function, and their parents, were asked to participate as the control group. Overall 18 families accepted. Additionally, in study II, a second group (control group 2) was used for comparison comprising 25 children with high and intermediate ARMs.

## 3.2 METHODOICAL CONSIDERATIONS

### 3.2.1 Study I

In the first study we translated and culturally adapted the disease specific quality of life questionnaire HAQL. The questionnaire was developed in the Netherlands for children and adolescents with fecal incontinence. The HAQL includes 10 or 11 domains, with questions concerning diet, laxatives, constipation, diarrhea, urine, and fecal incontinence, social and emotional functioning, body image, and physical symptoms. One domain concerning sexual function is included in the questionnaire targeting the oldest age group of 17 years and older, please see table 2.

**Table 2** presents HAQL domains and items within the domain. Two domains were excluded: questions concerning sexuality for the two older age-groups (SEF) (2 items). constipation (CON) (1 item).

DOMAINS	Number of items
Fecal continence* (FC)	8
Urinary continence (UC)	4
Social functioning (SF)	3
Emotional functioning*(EMF)	
Laxative diet (LD)	7
Constipation (CON)	2
Presence of diarrhea (PD)	2
Body Image (BOI)	2
Physical symptom* (PH)	2
	9

\*Evaluated domains

The questionnaire is available in three age-specific versions for the age groups (8– 11, 12 – 16 and 17+ years) and proxy versions for the two younger age groups [48]. The HAQL is also available in Italian, English, and French [59, 81, 82]. We obtained the English and the Dutch versions of the HAQL from the developers of the original study.

### 3.2.2 Study II

The second study concerned QoL and bowel function as well as an assessment of the translated and culturally adapted HAQL from study I. Seventeen children, (eight boys and nine girls), with a mean age of 11.2 years (range 7.4 – 18.3 years) and 18 mothers and 13 fathers of the 23 families who had accepted participation returned completed HAQL questionnaires. Table 1 shows domains and numbers of items within each domain. Each item consisted of two parts: 1. The respondent indicates how often a specific problem occurs over

a one-week timeframe, scored on a 4-point scale ranging from never to very often. 2. The respondent indicates how unpleasant it was, scored on a 4-point scale of not at all, a little bit, unpleasant, and very unpleasant.

Bowel function was assessed using a seven-item Bowel Function Score (BFS) [21, 30]. Twenty-seven families (61%) of 15 boys and 12 girls with a mean age of 14.0 years (range 8.0 – 22.7 years), returned completed BFS questionnaires. The BFS assesses voluntary control of defecation sensation, frequency, soiling, constipation, and social impact on the individual. Parents completed the score for children under 16 years of age. A total score of 17 – 20 is considered normal bowel function in a normal population [23]. The BFS is commonly used and has been validated for patients with ARM [83].

Sixteen families completed both the HAQL questionnaire and the BFS questionnaire. A control group comprising 17 children, and 16 mothers and 7 fathers of healthy children also completed the HAQL questionnaire.

### **3.2.3 Study III**

Study III concerned psychosocial function and, emotional and behavioral problems. To measure the children's and adolescents' psychosocial function, IAPSQ was used. Of the 23 families who had agreed to participate, 16 children and adolescents completed the IAPSQ, including seven boys and nine girls, with a mean age of 11.0 years (range 7.3 – 18.3 years). A control group comprising 17 healthy children also completed the IAPSQ. A second control group comprising 25 children with intermediate- and high ARM was used for comparison.

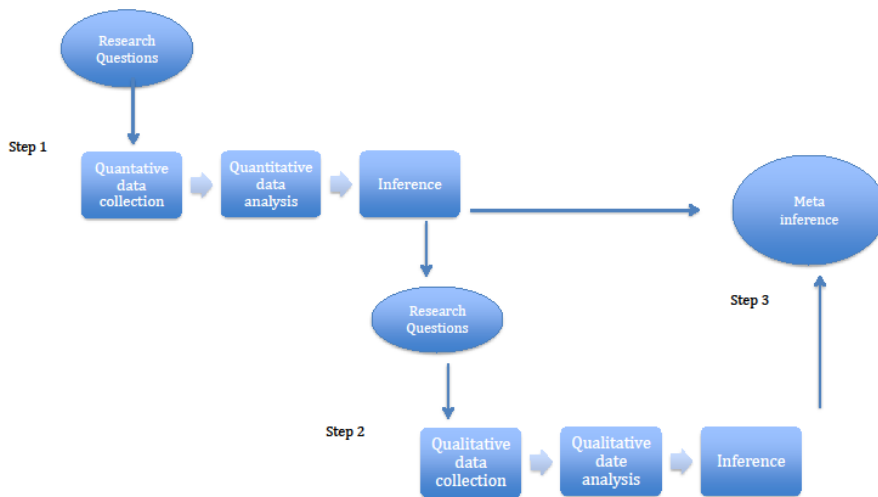
The IAPSQ is a validated questionnaire for children with ARM [68]. The IAPSQ was developed from a questionnaire for children with imperforate anus and was used in a previous study at Astrid Lindgren Children's Hospital [67]. The IAPSQ consists of 35 items. The items are classified into two domains: One social domain (SOC) and one psychological domain (PSYCH) including the three sub-domains; Emotional (EMO), Emotional/Cognition (COG), and Self-determination (SELF). The items are scored on a five-point Likert scale.

To measure the children's and adolescents' emotional and behavioral problems reported by their mothers, the Swedish version of the Child Behavior Checklist (CBCL/6-18) was used (Achenbach & Rescorla, 2001). Of the 23 families who had agreed to participate, 18 mothers

of nine boys and nine girls with a mean age of 11.1 years (range 7.3 – 18.3 years) completed the questionnaire. The control group consisting of 17 mothers of healthy children and adolescents also completed the CBCL. The CBCL contains a syndrome scale with 113 statements where the responses are rated as 0 = not true, 1 = somewhat or sometimes true, and 2 = very true or often true. Using a specific syndrome profile sheet, the answers are summed in subscales comprising items with problems that tend to occur together. The profile sheet reveals a T-score and presents the scores within clinical range, borderline range, or normal range. For the syndrome scale, a T-score <65 is considered to be normal, >70 is considered to be clinical, and in between is borderline. The subscales are summarized into a total score for, internalizing and externalizing behavior. Internalizing behavior comprises withdrawal, somatic complaints, and anxiety/depression. Externalizing behavior includes rule-breaking and aggressive behavior. The total score also comprises social problems, attention problems, cognitive problems and other problems.

#### **3.2.4 Study IV**

In study IV a sequential explanatory mixed method design was used. This method provided a structure for the collection of initial cross-sectional quantitative data through questionnaires, and it was followed sequentially by semi-structured interviews to add deeper understanding of the quantitative data. The interview questions were based on the initial quantitative findings. This design helped us to answer complex research questions concerning parenting stress in the context of low ARM. Figure 2 presents a flow chart of the sequential mixed method design.



**Fig 2** Graphic presentation of the sequential mixed methods design in which research questions of the second strand emerge from the findings of the first strand. In step 3 the integrated meta-inferences are made on the bases of both sets of findings

Parenting stress was measured using the SPSQ, which aims to measure parents' perception of stress in their role as a parent [80]. Of the 23 families who had agreed to participate, 15 mothers and 13 fathers of 18 children returned completed questionnaires. The group of children included 9 boys and 9 girls with a mean age of 11.2 years (range 7.3 – 18.2 years). The age group of 8 to 11 years included 14 children and four children, and the age group of 12 to 18 years included 4 children. The SPSQ contains 34 items scored on a 5-point Likert-type scale, where parents are asked to agree or disagree with each statement (1 = disagree to 5 = agree). The SPSQ is divided into the five subscales of incompetence regarding parenthood (INCOMPETENCE), restriction of roles (ROLER), social isolation (SOCISOL), spousal relationship strain (SPOUSE), and the parent's physical health (HEALTH). High values indicate high levels of stress. A total score was calculated as well as mean scores for each subscale.

All parents in the index group who had completed the questionnaire were asked to participate in an individual interview. Ten parents agreed, but one father who first agreed later withdrew his participation. Semi-structured interviews were conducted with nine parents, comprising two fathers and seven mothers. The interviews lasted for 30–90 minutes and were recorded and transcribed verbatim. The interview questions were derived from the results of the

SPSQ, and they focused on two of the subscales in the SPSQ where statistically significant differences were found, namely, INCOMPETENCE and SPOUSE

### **3.3 DATA ANALYSIS**

A p-value of <0.05 was considered statistically significant in studies II–IV.

#### **3.3.1 Study I**

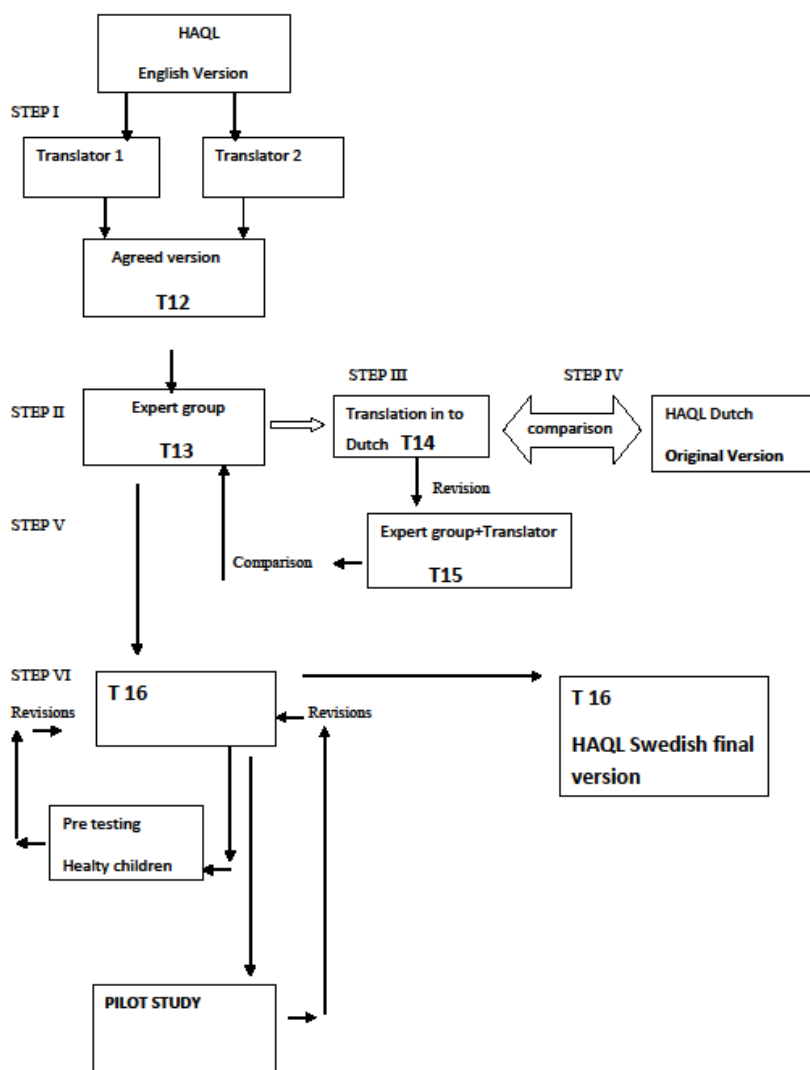
The translation and cultural adaption was initially carried out according to accepted guidelines and a forward/back translation method was used [49]. Alternations in the translation method were made during the translation process and an extra forward translation to Dutch was added. The translation process is explained step by step below and in figure 3. In the first step a forward translation was carried out by two independent translators from English to the target language Swedish. A consensus about the translation was reached and called T 12. In the second step an expert committee reviewed the T 12 version. The T12 was revised both linguistically and culturally. The revised versions was called T13. In the third step a native Dutch-speaking translator translated the T13 version into Dutch. The translated version was called T14.

In step four the Dutch translator worked together with two members from the committee. The translated Dutch version T14 was compared with the original Dutch version. Corrections were made in T14 and the new revised version in Dutch was named T15. In step five the T13 was revised by the Dutch translator and two members of the expert group, according to the adjustments in T15. The new Swedish version was called T16.

In step six the T16 was tested on healthy children and adolescents, two in each age group version of the questionnaire. The T16 had two additional questions about how they understood the questionnaire and if they wanted to add something. The questionnaires were reviewed after completion and minor revisions were made in T16.

A pilot study was carried out, and the T16 HAQL questionnaires were tested a second time on children with Hirschsprung's disease (HD). The questionnaires had the same two extra questions added as in the previous test. For the proxy versions there were extra lines added for "other comments". After completion, the questionnaires were reviewed.





**Figure 3** Flow chart over the translation process

### **3.3.2 Study II**

For the evaluation of the HAQL questionnaire, a Mokken scalability analysis [84] and a Rasch analysis [85] were carried out. For comparison between the child and the mother, the Wilcoxon matched pairs test or a paired t-test was used. For comparison of patients and the control group, the Mann-Whitney test for independent groups was used. In some domains where the data predominantly included only low or zero scores, the Fisher exact test was used. A Mokken scalability analysis and a Rasch approach were also applied for the BFS. The agreement with the HAQL was estimated using the non-parametric Spearman rank correlation. All data are presented as the mean and standard deviation (SD).

### **3.3.3 Study III**

Analyzing the IAPSQ questionnaire, both sum scores and scores imputed for missing values were calculated for each of the three sub-domains and the two domains. ANOVA was used to calculate the correlation of imputed sum scores between the index group, healthy controls, and the control group consisted of children with intermediate- and high ARM. Data are presented as means and SD. Adjusted (multiple) regression analysis was used to calculate differences between groups when adjusted for age and sex.

The CBCL questionnaire was analyzed according to the handbook for CBCL/6-18 (Achenbach & Rescorla, 2001). Total problem scores and groupings of internalizing and externalizing problems scores were first calculated and then converted into T-scores according to the provided profiles, where normal-, borderline-, and clinical ranges are presented. The T-test was used to analyze differences between the index group and control group for calculated T-scores, and the Mann-Whitney U-test was used to analyze the raw scores. Data are presented as means and SD.

### **3.3.4 Study IV**

#### *3.3.4.1 Quantitative analysis*

Descriptive statistics are presented as the number of observations (n), the mean (m), the SD, and the standard error of the mean (SE). Because the correlation between the mothers' and the fathers' scores was high, a linear mixed model was used between the groups (adjusted for parents' age and gender). One model for each subscale was used. The participants contributed with one or two observations depending on whether one or both parents participated. The

individual was treated as a random variable in the model, and all other variables were fixed. The model’s covariance structure was compound symmetry [86, 87] .

### 3.3.4.2 Qualitative data analysis

The transcribed text was analyzed using a qualitative content analysis method [88]. The interviews were first read through and interpreted by the first author and two of the co-authors. The text was then divided into meaning units condensed, and coded. The codes were first sorted into sub-categories and thereafter into categories, and this constituted the manifest content of the text. The categories were reflected on, and the latent contents of the categories were formulated into themes. (Table 3)

**Table 3** Examples of meaning unit, condensed meaning unit, codes, sub-category, category and theme

Meaning unit	Condensed meaning unit	Codes	Sub-category	Category	Theme
I don't think you can understand how it's going to be a parent before becoming one, you have some thoughts about it but it was so much larger than one can ever understand	To become a parent, it's so much larger than you ever can understand before becoming one.	To come to insight and gain experiences	Insights and reflections Hard to comprehend before becoming a parent	Experiences	Expectations of parenthood
You don't really understand, because you don't know	Can't comprehend before, because you don't know how it is	Insight			

### 3.3.4.3 Integration of quantitative and qualitative data

In this mixed method study, qualitative findings were used to explain and expand the quantitative findings. Inferences were made through integration with comparison, correlation and consolidation of the two types of data. The two strands of inquiry were then merged. The process is explained in figure 2 [89].

### **3.4 ETHICAL CONSIDERATIONS**

The study was approved by the Ethics Review Board in Stockholm, Dnr 2008/1706-313 and was carried out according to the Helsinki Declaration [90]. Research involving minors is ethically challenging due to children's limited ability for risk- and consequence assessment. The principle of autonomy is put aside in the field of pediatric research when the parents as legal guardians play the role as advocates for their children. In order to respect autonomy and the ability to decide for themselves, it is important to explain the purpose of the study in an understandable way for children using age-appropriate information. This is to enable the child to comprehend the information presented, and with that said, all participants including both parents and children received written information prior to the study. The information was age adjusted for the children. All participants were informed that participation was voluntary and that they could withdraw their participation at any time. They also provided written informed consent prior to participation. The participants were also informed that all material would be treated confidentially. Interviews were performed with parents. Before the interviews the interviewer was assured that the informant agreed to recording and transcription of the interview.

## **4 RESULTS**

### **4.1 STUDY I**

When version T14 was compared to the original HAQL in Dutch, it was discovered that Question 29 had been altered from its original meaning. In T14 the children were asked if there had been feces in the bowel without an urge to defecate. In the original Dutch HAQL the children were asked if they went to the toilet without the urge to defecate. Question 29 was corrected in T15. The English HAQL had been translated but not culturally adapted [81].

The T15 version was overall very similar to the original HAQL instrument in Dutch. In many of the questions the T15 was almost identical to the original HAQL version in Dutch. In some of the questions the words differed from the original, but the true meaning of the words were the same. In the original Dutch version, the language was more simplified. We chose to use the more simplified language in the questionnaires intended for the children but used another choice of words in the versions for proxies and adolescents.

The T16 version was first pre-tested on six healthy children/adolescents, two in each age group. The children had difficulties understanding the word translated as condition (åkomma) and the word was changed into the word inconvenience (besvär).

No specific question stood out in the pilot test. Some of the participants had difficulty understanding the content in isolated questions. The questions concerned differed, and there was no pattern, and there was no question that recurred among the participants. One family had problems with one question and chose not to complete that particular question. Examples of received comments were “too many questions”, “tedious”, and “strange”. Others, especially the younger participants, experienced the form as a little too long. The outcome of the translation process of the HAQL resulted in a final version, (T17), of the Swedish HAQL.

## **4.2 STUDY II**

### **4.2.1 Outcome of the HAQL**

Compared to the healthy control group, the children and adolescents with low ARM reported significantly lower function in the Physical symptom (PH) ( $p = 0.045$ ), Fecal continence (FC) ( $p < 0.01$ ), and Laxative diet (LD) ( $p < 0.01$ ) domains. Differences were also found in the Emotional functioning (EMF) domain, in which children with ARM scored lower functioning, although the result was not significant. No systematic differences could be statistically demonstrated in the other domains.

Children and adolescents with low ARM and their mothers were matched on a pair level. The children scored significantly lower function and more symptoms than their mothers in the Emotional functioning (EMF) ( $p < 0.01$ ) and Physical symptom (PH) ( $p = 0.018$ ) domains. No systematic differences were found between the scoring of the mothers and children in the other domains.

Both the mothers and fathers were asked to complete the questionnaire in the control group as well as in the index group. Compared to the mothers' response rate, the response rate among the fathers, particularly in the control group (six fathers,) was low. Because the answers among the participating fathers were similar to the mothers' answers, the fathers were not included in the analysis. The domains concerning Sexual function (SEF) and Constipation (CON) were not analyzed.

In the FC, EMF and UC domains, children from the younger age group (index group 8 – 11 years) reported more symptoms than those of the older patients (index group 12 – 16 years) but, the differences were not significant.

#### **4.2.2 Outcome of the BFS**

Twenty-seven families (62.7 %) completed the BFS (Table 1). The overall mean score was 17.2(1.9), which is comparable to the Finnish population with low ARM [18]. Seventeen of the patients (63%) scored  $\geq 17$ , which is within the normal range, and 6 of these (22%) scored 20. Nine (33%) of the patients scored between 12 and 16, which is a moderate outcome, and one patient (4%) scored 10. The 15 boys scored a slightly higher mean of 17.7 (2.8) than the 12 girls, who had a mean of 16.3 (2.0). The younger patients (8 – 11 years) scored lower with a mean of 16.3 (1.9) compared to the older patients with a mean score of 17.0 (1.6). None of these differences were significant.

#### **4.2.3 Evaluation of the HAQL**

Due to the low number of participants (17) in the index group we were not able to verify or question the different domains in the questionnaire. However, a parsimonious evaluation of the HAQL domains FC, EMF and PH using a Mokken scalability analysis and Rasch analysis showed no obvious conflicts in any of the investigated domains. It should be noted that in most of the matched pair reports, both children and mothers reported “never” (0) when answering and a few mothers reported “I don’t know”.

A comparison between the BFS and HAQL was also performed. Sixteen pairs were used in the comparison. No correlation was found between the BFS and the HAQL domains FC and PH.

#### **4.2.4 Drop-out analysis**

Using descriptive statistics, all patients with ARM who participated in the study were compared to non-responding patients using hospital records. The groups were similar regarding the patient’s bowel function when comparing the non-participants with ARM and participants with ARM at the most recent check-up.

### 4.3 STUDY III

#### 4.3.1 Outcome of the IAPSQ

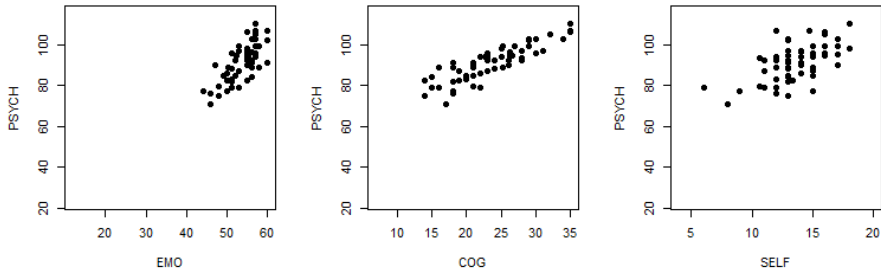
Sixteen (36%) children and adolescents with low ARM answered the IAPSQ. A control group consisting of 17 healthy children also answered the IAPSQ. A second control group consisting of 25 children with intermediate- and high ARM was used for comparison. The children and adolescents with low ARM scored high overall in the psychosocial domains. Compared to healthy controls, similar scores were found in all domains. When comparing the index group with the control group of children with high- and intermediate ARM, no statistical differences were found, except for in the domain COG, where the reference group scored significantly lower function ( $p = 0.042$ ), see table 4.

**Table 4** Sum score comparison in groups: sub domains emotional/cognition (COG) emotional (EMO) self-determination (SELF) and the domains psychological (PSYCH) and social (SOC) presented as mean and standard deviation (SD).

	Index Group n= 16	Comparison Group 1 n=17	Comparison Group 2 n=25	p-value
COG	25.55 (4.86)	25.13 (6.08)	21.61 (5.33)	0.042*
EMO	54.76 (3.13)	53.26 (3.78)	53.54 (3.97)	0.460
SELF	14.25 (2.41)	13.82 (2.51)	13.11 (2.16)	0.295
PSYCH	94.56 (8.47)	92.21 (9.96)	88.25 (8.10)	0.076
SOC	51.24 (4.47)	49.36 (6.37)	49.86 (4.89)	0.567

When adjusted for age and sex, the difference regarding the domain COG still had significantly higher scores ( $p=0.03$ ) for the index group compared to the second control group. A calculated sum score of the domain PSYCH (sub domains EMO, COG, and SELF) also showed significantly higher scores for the index group ( $p = 0.02$ ) compared to control group. There were no correlations found between the different domains and the child/adolescent's age or sex.

Calculated correlations regarding the three groups between the three sub domains EMO, COG, and SELF and the sum score of the domain PSYCH showed that the sub domains with the highest covariance were COG ( $\rho = 0.88$ ) and EMO ( $\rho = 0.70$ ) and the sub domain with the lowest covariance was SELF ( $\rho = 0.54$ ), see figure 4. No major differences were found between sum scores and imputed values. Missing values were found mostly in the healthy control group.



**Figure 4.** shows covariance in the sub domains Emotional (EMO), Emotional/cognition (COG) and Self-determination (SELF) and sum score of the domain Psychological (PSYCH).

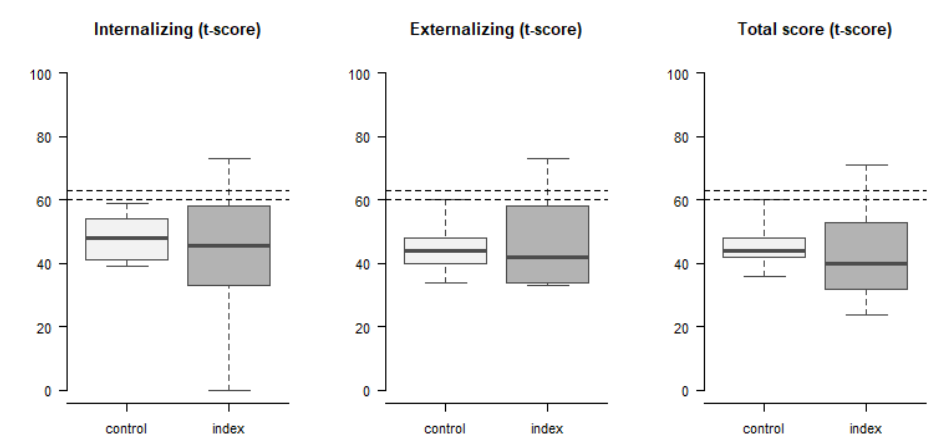
#### 4.3.2 Outcome of the CBCL

Thirteen (72%) of the children and adolescents with low ARM were scored by their mothers, within normal range of the syndrome scale. In the total problem score, three children were scored for clinical behavior problems and none for borderline problems. Three children had scores compatible with internalizing behavioral problems within the clinical range, and none were within borderline range. A total of four children had scores compatible with externalizing behavior problems, three within the borderline range and one considered to be within the clinical range. Ninety-four of the children in the healthy control group were scored within the normal range on the total syndrome scale, and one child was reported within the borderline range for total problem. One child was scored with externalizing behavior problems and none of the children in the control group were scored with internalizing behavioral problems. No significant differences were found between the index group and the control group on the syndrome scale. The distribution was larger in the index group compared with the control group. For descriptive data, see table 5 and figure 5.



**Table 5** CBCL comparisons between children with low ARM and comparison group 1. Numbers of patients in each group and percentage within brackets.

	Index group n=18	Comparison group 1 n=17
<b>Syndrome scale</b>		
<b>Internalizing</b>		
Normal	15 (83)	17 (100)
Borderline	0	0
Clinical	3 (17)	0
<b>Externalizing</b>		
Normal	14 (78)	16 (94)
Borderline	3 (17)	1 (6)
Clinical	1 (6)	0
<b>Total problem</b>		
Normal	15 (83)	16 (94)
Borderline	0	1 (6)
Clinical	3 (17)	0



**Figure 5.** Boxplot comparison of CBCL syndrome scales. T-scores regarding externalizing and internalizing behavior and total in index group (index) and comparison group 1 (control). The thick line represents the median, the box represents the interquartile range, the bars represent the upper- and lower quartiles. The values below the two dashed lines are normal range, above are clinical-, and in between borderline range.

## 4.4 STUDY IV

### 4.4.1 Quantitative results

#### 4.4.1.1 Outcome of the SPSQ

The total mean score of all five subscales in the questionnaire for the parents in the index group was  $m = 2.29$  ( $SD\ 0.48$ ). Mean scores for the subscales were INCOMPETENCE  $m = 2.02$  ( $SD\ 0.51$ ), ROLER  $m = 2.96$  ( $SD\ 0.70$ ), HEALTH  $m = 2.38$  ( $SD\ 0.69$ ), SPOUSE  $m = 2.14$  ( $SD\ 0.65$ ), and SOCISOL  $m = 2.08$  ( $SD\ 0.73$ ).

No significant differences were found between parents in the index group and healthy controls in the total score or in any of the subscales except for the subscale INCOMPETENCE where the control parents scored significantly higher levels of stress compared to the parents in the index group ( $p = 0.003$ ). Slightly higher stress levels for the control group were also found in the ROLER and SPOUSE subscales, but these differences were not statistically significant.

The variable most likely to explain stress was the child's sex. Parents of girls showed significantly higher stress levels compared to parents of boys for the total score ( $p = 0.016$ ) and the subscales INCOMPETENCE ( $p = 0.033$ ) and SPOUSE ( $p = 0.023$ ).

No differences were found between mothers' and fathers' reports or when comparing younger (8–11 years) and older (12–18 years) children in any of the subscales or in the total score.

### 4.4.2 Qualitative results

The results from the interviews resulted in three themes – *Communication between parents*, *Expectations of parenthood*, and *Challenges concerning parenthood*.

#### 4.4.2.1 Communication between parents

Different approaches to communication were presented in this theme, and parents described how they cooperated and how they dealt with the responsibilities of parenthood. A well-functioning cooperation with the other parent and a shared responsibility for parenting was described as well as different roles within the relationship and that the roles could shift depending on who was on parental leave. “*To pull together*” was a recurrent term. A more traditional role of parenting, where the mother has the primary responsibility and the father helped was also represented in the theme. Several of the parents also talked about the

importance of being responsive in the relationship and, to listen to one's partner and be able to adapt to the situation.

#### *4.4.2.2 Expectations of parenthood*

Many of the parents talked about how, after having children, they had gained experience and come to realize what it means to be a parent as well as the difficulty in understanding what it meant to be a parent before becoming one. Past events in a person's life, both good and bad, might come to characterize how a person performs their role as a parent, and experiences such as previous pregnancies, older siblings, and traumatic

births all have an impact on a person's parenthood. One parent told how previous traumatic births led to anxiety and great concern during the pregnancy. One mother had, because of her own bad experiences in childhood, thought a lot about what kind of parent she wanted to be before becoming one.

When the child was born and during the difficult periods with operations the feelings of being here and now in the middle of chaos were described, and several parents talked about not being able to plan for more than a short period at a time. "Here and now" and "taking one day at the time" were recurrent statements in this category.

When parents reflected on being a parent to a child with low ARM, positive thoughts and the belief that everything is going to be fine were described. One father told how his expectations on becoming a parent corresponded well with the reality of being a parent despite the malformation.

#### *4.4.2.3 Challenges concerning parenthood*

Positive statements about being a good parent and being satisfied with themselves as parents were given. They had reflected on themselves as parents and stated that everything had turned out just fine. One mother stated that she was satisfied with the way she was as a parent but emphasized that she was good as a parent together with her partner.

Expressions of inadequacy and feelings of guilt towards siblings were also expressed. Parents compared the siblings to each other when they reflected over being a parent. Several parents also described how they treated the siblings differently. In some cases, the fact that the child was born with a malformation affected the way the parents acted. One mother found that the child had been more cherished than the sibling, partly because the child was the first born and partly because of her low ARM. Another mother noted that it was more important to have routines and provide information for her girls than for her son (both girls were born with low

ARM). Other parents stated that they treated the siblings differently, but it had more to do with the child's personality or if the child was the oldest or youngest among the siblings, rather than the low ARM.

## 5 DISCUSSION

The overall aim of this thesis was to investigate how children with low ARM and their parents are affected by the malformation. The thesis provides us with a holistic approach to the task and various aspects have been compiled to obtain as complete a picture as possible of the consequences the malformation may entail. We approached the task by investigating QoL (*Study II*), psychosocial aspects (*Study III*), and physical function (*Study II*). Parents' experience of parenting stress related to the malformation was also investigated (*Study IV*). Unfortunately, we were not able to investigate the siblings' QoL as was originally intended. Due to low response (only eight siblings) we had to abandon that approach, although the parents did talk about different aspects of the siblings in paper IV. The different aspects of this thesis will hereby be further discussed.

### 5.1 MAIN FINDINGS

#### 5.1.1 HAQL

In study I we aimed to translate and culturally adapt the disease-specific questionnaire HAQL into Swedish. An evaluation of the translated questionnaire was also performed (*Study II*). The reason why we took on this task was the lack of disease-specific QoL questionnaires available in Swedish for children and adolescents with ARM. To assess disease-specific QoL in patients, multidimensional self-report questionnaires are required.

When an individual's health status is assessed, the concept of HRQoL is used. HRQoL focuses on health care aspects [40] and is defined as a multidimensional concept that covers social, physical, and psychological health domains [34, 91, 92].

The choice of a disease-specific QoL instrument for our purpose seemed most relevant, because difficulties with fecal impairment might limit the child in daily life activities. Compared to generic instruments, disease-specific questionnaires are considered to be more sensitive and to focus on specific problems of a particular disease [40]. Generic instruments, on the other hand, are intended for a more general use and allow comparison between different conditions and populations. Further on, the terms QoL and HRQoL will be used interchangeably.

A questionnaire is designed and validated for a specific setting and language, and in order to be used in other settings a translation and cultural adjustment must be implemented. For the translation we chose to use a modified process that considered the challenges of a small sample size in a rare disorder. The applied version was simplified and altered from the published guidelines [49, 54]. The recommendation in these guidelines is to use large sample sizes and numerous professional translators, which was an expensive and time-consuming process. However, Acquadro et al. [53] concluded that there is no evidence in favor of a certain method of translation and recommend a multistep approach to guarantee quality of the translation. Price and colleagues presented an example of a modified and simpler way to approach the problem, which was a cross-cultural translation method for rare pediatric populations [93].

During the process of translation questions arose concerning the translation of the English version of the HAQL. We therefore decided to alter the translation process with a translation to ensure the accuracy of the translation. A back translation is intended as a quality control to ensure that the translation has the same meaning after translation [52].

We therefore performed a second translation into Dutch, the original language and compared the two Dutch versions. The comparison confirmed the lack of cultural adaption of the English translation. It was also discovered that one question had been altered from its original meaning. This points out the importance of a second translation as a quality control [49]. The additional comparison between the Swedish version of the HAQL and the original Dutch version verified the correspondence between the two questionnaires.

An evaluation of the HAQL was performed in Study II. However, the sample size was too small to perform a complete verification of the domains in the HAQL. Instead we performed an investigation of the item-set in each domain. The reason for this was to establish how the questions correlated and contributed to the domain and to see if the questions fit within the domain. An additional problem was that the number of items within each domain varied from 2 to 9 and, dimensions with fewer than three items are more likely to be unstable [94]. Three domains: fecal continence, emotional function and physical function were evaluated on a basic level. No obvious conflicts were found, and the questions seemed to function as intended. Due to non-response to some of the items, a comparison of sum scores would affect the analyses. Therefore, a transformation to a metric measure via a Rasch approach was performed for the three domains. Further, the Rasch model enabled us to include incomplete questionnaires opposed to the Mokken analysis, which may be problematic with a small

sample size. To obtain as much information as possible for the validation, the mothers and children's questionnaires were combined into a set of 34 questionnaires.

Several of the domains of the original structure of the HAQL were found to be weak in its original form [48]. A likely explanation was the small item set with only two items in some domains. To obtain a valid, and for the children shorter and easier questionnaire, and to achieve a more solid structure for the adolescents and adult HAQL versions, a modified structure with fewer dimensions containing only two items, has been suggested by the developers of the French version [95]. A structure with fewer items and domains for children and proxies was also recommended by the French research team [82].

### **5.1.2 Outcome of the children and adolescents**

#### *5.1.2.1 QoL and functional- and psychosocial outcome*

In study II we aimed to investigate QoL and bowel function. The core findings in study II indicated impaired QoL among the children and adolescents in some of the HAQL domains. Overall, they appeared to have more physical problems and fecal incontinence compared to their healthy peers. We also found differences between the two groups in emotional functioning where the index group had impaired function, even if the results were not significant. Differences in body image and emotional functioning have been considered to be the most important factors concerning the patients' QoL [96]. Absence of fecal problems has been considered to be an important predictor of good QoL in patients with ARM. Correlation between impaired bowel function and QoL has been reported in previous research and fecal continence was found to be the most relevant and negative factor affecting HRQoL in patients with ARM [97]. Further, mental well-being and QoL were affected by problems such as constipation and fecal leakage in adolescents with ARM or with chronic idiopathic constipation [98]. In order to achieve better QoL, it was found to be crucial to handle problems such as fecal soiling [76]. However, there are also studies suggesting the opposite where no correlation was found between problems with fecal incontinence and reported QoL [62, 99].

The BFS evaluation of the long-term functional outcome in the index group showed similar results as were found in the Finnish population with low ARM [11, 23]. The Finnish study showed impaired bowel function in patients with low ARM compared to healthy controls. Unfortunately, we did not have a comparison group when conducting our study. However,

the similar results and the similarities between Sweden and Finland give us an indication of the bowel function in a Swedish population with low ARM.

When compared, we did not find any correlation between the HAQL domains regarding bowel function and the BFS. An analysis of fecal incontinence scores performed by Bischoff et al. concluded that scoring systems, especially those designed to measure QoL are inadequate because of their subjective assessments and are of little use in handling pediatric patients [100].

However, we found similar results concerning bowel function and age in the two questionnaires. The younger age group (aged 8 to 11 years) seemed to have more impaired bowel function than the adolescents (aged 12 to 16 years) in both questionnaires. The BFS evaluation had an additional group (aged 17 to 23 years) that scored even better function than the two younger age groups. Our results confirm previous studies, where children with ARM had more fecal problems than adolescents [57, 99]. Danielson and colleagues found that adult patients with perineal fistulas (low ARM) did not differ much from a control group even though many of the patients had suboptimal bowel function. [101].

Few studies have been conducted on psychosocial function in ARM patients, but this area was to some extent studied during the 1990s and early 2000s. In study III we aimed to examine psychosocial function and possible emotional and behavioral problems in the study group. An additional control group consisting of children with high and intermediate ARMs was also used in study III, which enabled us to examine correlations between psychosocial function and the severity of the malformations. The main finding in study III was that the children and adolescents have good psychosocial function. They did not differ from the control group consisting of healthy children, and they scored good function in all of the IAPSQ domains. Compared to the children with intermediate and high ARM (control group 2), the index group children had similar results except for in one domain (COG), where the index group had better function. We also found differences with higher scores for the index group compared to control group 2, when a sum score was calculated (including the EMO, COG, and SELF domains) for the PSYCH domain. The severity of the malformations and problems with fecal incontinence and constipation that follow can to some extent explain our results. The index group consisted of patients with milder forms of ARM compared to control group 2. When compared to the milder forms, the function among the more severe types are known to be impaired. Functional impairment appears to correlate with the severity of the malformation, and patients with more severe forms, also reported impaired bowel function



[56, 96]. Our findings also support previous results showing that poor psychosocial function among adolescents with ARM is associated with poor fecal continence [102].

In contrast, no correlation was found between functional problems and psychosocial function in children with ARM, except for in girls aged 6 to 11 years with incontinence [103].

Furthermore, previous results from the IAPSQ did not show any psychosocial impairment in children with intermediate and high ARM despite problems with fecal incontinence reported by their parents [104]. One explanation might be differences in analysis methods. We analyzed the results in grouping by domains, while the earlier results [68] were analyzed question by question.

Our results in studies II and III confirm to some extent the findings in previous studies, Hartman and colleagues reported impaired physical, psychosocial, and overall QoL function among adult ARM patients when compared to controls [60], and patients with ARM had more problems with constipation, soiling, and gas incontinence as well as impaired QoL when compared to controls [17].

#### 5.1.2.2 Proxy evaluation

The children and their mothers had similar outcome on the HAQL except for in two domains where the children scored more symptoms and lower emotional functioning (*study II*). It is well established that child self-reports should be considered standard when measuring HRQoL in children [39] and that children's self-report is not synonymous with proxy-reported assessments [40].

Even though self-report instruments should be considered standard when measuring QoL, a child could be too young or too sick to complete a self-report, and a parent-proxy report might be needed [39]. It is important with proxy informants in pediatric care, where patients' due to their developmental stage, ability to read, or language skills might be considered to be unreliable [81]. A combination of both proxy tools and child self-reports was also found to be complementary and relevant [82].

In contrast to our findings in study II, in children with chronic conditions, parents tend to report lower QoL than the children [105, 106]. Few studies have been conducted on comparison between self-reports and proxy reports in children with ARM and the results between conducted studies vary. Parents of children with ARM overestimated both physical- and emotional functioning, when generic QoL was measured [45]. On the other hand, when

psychosocial aspects in children with high and intermediate ARM were examined, there were discrepancies between mother's assessment and children's self-reports. The mothers considered the children to be sadder and angrier than the children themselves [43].

Such a comparison as in Study II could not be performed in Study III, where different instruments were used for children and parents. However, the results tend to point in the same direction with discrepancies between proxy- and children's self-reports answers. Even if the children and adolescents scored good psychosocial function, the results from the CBCL scored by their mothers, suggest that they might have emotional and behavioral problems. Five children, (33%) were scored within clinical or borderline range in one or several of the problems scores on the syndrome scale. Only one child (6%) had borderline externalizing behavior problem in the healthy control group.

More emotional and psychosocial problems were also found in a CBCL evaluation in children with high and intermediate ARM, esophageal atresia and congenital diaphragmic hernia, when compared to the general population [107]. Furthermore, children from the same study population who underwent multiple surgery and had home medical treatment also had higher T-scores compared to children who underwent single surgery and did not have home medical treatment [108].

Upon examination of research data, the parents' well-being emerged clearly during the interviews. Further when comparing individual interviews and family responses from the surveys, there were indications that individual parents' well being influenced their proxy rating as there were striking differences in, not just parent – child but also parent – parent answers. There are several factors to consider that may affect the level of agreement between parent and child, for example: parents well-being, socio economic status, family structure and the severity of the condition in the child [109, 110].

Mother's depression was found to negatively correlate with proxy reports of HRQoL [111]. Furthermore, a risk factor found to correlate negatively with children's QoL, were maternal posttraumatic stress syndrome [108]. To conclude, assessment of QoL in pediatrics should not only include child self-reports and parent proxy reports. The family- and social context should also be considered [112].

### 5.1.3 Parenting stress

In study IV, parenting stress was explored. We chose to immerse ourselves in the topic using a mixed method containing both a quantitative and qualitative approach, as well as merging of the results from the two strands. The quantitative results did not reveal any major differences when parents in the index group were compared to parents of healthy children. Only one subscale, INCOMPETENCE showed a significant difference, where the parents of children with low ARM scored lower levels of stress. This was an unexpected result because previous research mainly focused on the negative aspects and increased parenting stress levels related to having a child with a chronic condition [73, 113]. Interviews with parents in the index group provided additional explanatory evidence with more details and life contexts. Although the quantitative findings did not reveal any high levels of stress, the interviews provided a more in-depth knowledge into earlier experiences of difficulties and stress, but also positive statements about parenthood. This is a good example of, according to Creswell and Plano Clark, a situation where the qualitative data aim to help us better understand the results from the quantitative findings [89]

Difficult experiences of parenting a child with low ARM were described, but also how these experiences might be the reason for why the parents seemed competent in their parenting role, as they appear to have grown and become stronger. We also found that parents in a strong relationship expressed the importance of cooperation and responsiveness and the importance of listening to one's spouse/partner.

Good family function and the family working closely together have been shown to play a central role for the healthcare provider's own health [114]. Further, better family functioning was found to correlate with reduced parenting stress [115]. Terms such as "*teamwork*" and "*to pull together*" were mentioned as important when going through a difficult period. Our findings support previous results where it was shown that parenting stress might activate problem-focused and emotion-focused coping behaviors [116].

To find out that one's child was born with low ARM was described as feeling as if the world was turned upside down. Studies regarding parenting stress related to their child's chronic condition present similar experiences with feelings of being overwhelmed and increasing stress after receiving the diagnosis [117, 118]. Feelings of inadequacy, only being able to take one day at a time, and the uncertainty they felt about the future was also brought up during the interviews, which is consistent with previous findings where reported parental care is related to information about the child's disease, emotional stress, feelings of guilt, and

uncertainty about their child's future health care needs [119]. Other identified sources of parenting stress are uncertainty about the child's diagnosis and lack of predictability [120].

The parents reflected positively on their parenthood and said that everything had turned out just fine, although feelings of guilt and how things could have been done differently – were present. This might help us understand why the parents reported significantly lower stress levels in the INCOMPETENCE subscale. They might have grown as parents because of what they have been forced to go through and might have adapted to their new context. Positive findings such as personal growth were also discovered in an investigation of psychosocial experiences of parents of children born with ARM [78]. Hastings et al discovered positive perceptions such as personal growth among parents of children with disabilities [121]. Further, Trute and colleagues found positive transformation to be a potential outcome when family strength was investigated in families of children with disabilities [122].

Feelings of guilt concerning siblings, and the fact that the siblings had been treated differently were also revealed during the interviews. The findings relate to previous observations where difficult experiences such as stress associated with ARM were found to affect all family members [77]. Others have also found parental feelings of guilt to be a cause of parenting stress [123].

Despite the fact that low ARM is considered mild, parents described the chaos they felt in having a child with a malformation and feelings of uncertainty about the future, as well as feelings of uncertainty about the future during periods of difficult procedures and operations.

Our results might be explained by the fact that the children and adolescents were older when the questionnaire was completed. The results might have turned out different if the questionnaire had been completed when the child was younger or newborn. Parenting stress in parents of children with a chronic disease appears to decrease over time [120] [124]. Furthermore, when parents of newborns with ARM were compared to parents of older children with the same malformation, the researchers found higher levels of anxiety amongst the parents of the newborns [75].

## 5.2 METHODOLOGICAL CONSIDERATIONS

The studies presented in this thesis contains both quantitative and qualitative methods. The variety of methods helps to provide in-depth knowledge of the research topic. Research data were obtained through various questionnaires (*Studies II-IV*) and interviews (*Study IV*). A translation and cultural adaptation of a disease-specific questionnaire (*Study I*) as well as evaluation of the questionnaire (*Study II*) was also executed. A mixed methods approach was added as well (*Study IV*). The aim for the qualitative strand of the mixed method study was to add more in-depth knowledge and obtain a more detailed understanding of parenting a child with low ARM and the stress it might bring. A mixed methods design helps to bring different perspectives and methodological expertise into the data analyses [89].

The translation process (*Study I*) is stringent and time consuming but ultimately the uncertainty about all of the work results in a reliable product. However, it is necessary to obtain a reliable instrument. A poorly executed translation might result in an instrument that is not comparable to the original [54]. There are several translation methods described and discussed in the literature [49-53], even if recent research indicates the use of back translation might not be mandatory [55]. Consensus concerning the best method has not yet been established. The cultural adaption of an instrument clarifies that the underlying concepts of the adapted questionnaire are those of the original instrument [125]. The translation process and the evaluation of the HAQL were executed in two steps (*Study I* and *Study II*). Epstein et al emphasize that the adaption and validation of questionnaires are two separate processes that should be distinguished [55].

The evaluation of the HAQL domains (*Study II*) showed no evident dispute in the domains investigated however, for a proper validation larger sample sizes are needed. Also, the analysis indicated a problem of the questionnaire to capture the target population, because they appeared to be “*too healthy*” for the questionnaire, and to verify a lack of conflict within the HAQL domains, a larger population covering the entire spectrum is needed. Further, the instrument is developed for two different congenital malformations, ARM and HD, and even if the two diagnoses have similar outcomes, there are also differences. Questions relevant for one diagnosis might be not only be irrelevant, but also difficult for the other population. This will also affect the accuracy of the questionnaire.

Other questionnaires included in *Studies III* and *IV* are considered reliable and are validated. The SPSQ is valid for parents with children 0 to 12 years of age, but the children in this study ranged between 8 and 18 years of age. However, there were no differences when younger and older age groups were compared. Also, for the BFS questionnaire, there was no comparison

group. However, the similarities between the two countries (Sweden and Finland) and the similar results could give an indication of the bowel function using the BFS in Swedish children with low ARM.

The thesis' main limitation was the sample size. Despite the fact that all available children and adolescents with low ARM who also met the criteria for this study were asked to participate. We also extended the collection for the research data twice in an attempt to improve the sample size. Further, our hospital is one of the largest referral pediatric hospitals in Sweden however, a national collaboration in order to improve the sample size would have been beneficial. Low response rate also affected the sample size. Several of the non-responding families were lost to follow-up, and others declined participation with the response that their child did not have any problems with their bowel function. A higher participation was seen in the BFS in *Study II* (62.7 %). Factors that could have affected the positive participation were that the questionnaire was included with the invitation to participate and that there were only seven questions to answer. Strategies recommended to improve postal survey response rates are for example, shorter questionnaires [126]. This is a delicate problem as mentioned earlier, where domains within questionnaires require a certain amount of questions in order to be accurate. However, the drop-out analysis performed in *Study II* did not reveal any differences between the responding patients and the non-participants concerning documented bowel function recorded in the medical charts from the most recent follow-up. For that reason, we do believe that the outcome has not been affected by the drop out to any substantial extent. This study refers to the milder forms of ARM, and the participation might have been different if the whole spectrum of the malformation had been included. A much higher participation was seen in a prior study on children with intermediate and high ARM performed at our center [104].

Additional limitations were that we did not obtain parental socio-economic and educational profiles because these factors are known to affect parental stress. Moreover, the cross-sectional design limited the ability to understand how the children and parents report QoL, functional- and psychosocial outcome as well as parental stress over time.

## **6 CONCLUSIONS**

This thesis has contributed to a deeper understanding of how children/adolescents with low ARMs and their parents are affected by the malformation. It is also an attempt at a holistic approach with both quantitative and qualitative methods to address this challenging task.

The overall findings from the four studies can be concluded as follows:

### **Study I**

The translation and cultural adaption of the HAQL instrument provides us with a disease specific QoL instrument for children and adolescents with HD and ARM.

### **Study II**

The children and adolescents with low ARM had good QoL even if they seem to have impaired bowel function and worse emotional functioning compared to healthy control patients. The mothers underestimated their children's physical symptoms and overestimated their emotional functioning.

### **Study III**

Children and adolescents with low ARM had good psychosocial function although there were signs of emotional and behavioral problems in some of the children. The severity of the malformation might be related to psychosocial function.

### **Study IV**

The parents of children with low ARM had low levels of stress. They appeared to do well and seemed competent in their parenting role. However, when interviewed the parents told about earlier experiences of guilt and stress from the time around the child's birth as well as the time associated with operations and other procedures.

## 7 CLINICAL IMPLICATIONS AND FUTURE RESEARCH

This thesis has contributed to increasing our knowledge on how an individual and the family can be affected by a rare congenital malformation. In addition, the thesis has also contributed to providing a disease-specific QoL questionnaire for this specific population. It is important to continue to follow these patients and their families over time, in order to obtain a deeper understanding of the impact of the malformation and how it affects the entire family.

Investigating the siblings' QoL and how the siblings might be affected would be a future aim as this is an important part of the family constellation.

The findings also provide us with knowledge on the parental experience of having a child with low ARM and illuminate some of the challenges that parents encounter. This knowledge can help us to better understand the stress that parents face in their role as parents to a child with a congenital malformation. In order to provide appropriate resources, the healthcare provider must assess each family's specific needs, and the impact that the malformation might have on the whole family. The parents require disease-specific nursing care, including recurrent information, education, close follow-ups, and support in everyday life.

Healthcare professionals who are trained in this field can, from the results in this study, obtain a deeper understanding of the malformation and its consequences. To be able to provide good quality care, it is essential to have such competence. To reach this competence in the field of nursing, it is required to have proper training and experience, which is a challenging task in today's health care system. This must be prioritized to be able to offer good follow up and support to families, children, and adolescents with this diagnosis.





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## Appendix

## HAQL FRÅGEFORMULÄR

Frågorna handlar om hur du handskas med dina besvär. Analatresi, (då har du inget bajshål) eller Hirschsprungs sjukdom (det är en tarmsjukdom). Frågorna handlar också om hur du hanterar dina besvär.

Frågorna handlar om de senaste 7 dygnen. Du får bara välja ett svar per fråga. Välj det svar som passar bäst. Det finns inga svar som är rätt eller fel, det är viktigt vad du tycker. Försök att svara på varje fråga och hoppa inte över någon fråga.

## Exempel:

Fråga: Hur ofta har du haft löst bajs under de senaste 7 dygnen?

Om du inte haft löst bajs under de senaste 7 dygnen ringar du in aldrig

Så här:

Aldrig Ibland Ofta mycket ofta

Om du ringar in aldrig, så kan du gå vidare till nästa fråga.

Om du haft löst bajs, **ibland**, **ofta**, eller **mycket ofta**, så ringar du in ett av de här svaren.

som här: aldrig **ibland** ofta mycket ofta

eller så här: aldrig ibland **ofta** mycket ofta

eller så här: aldrig ibland ofta **mycket ofta**

Under **ibland**, **ofta** och **väldigt ofta** kommer du att se frågan "Hur jobbigt tyckte du att det var"

Kryssa i hur jobbigt du tyckte att det var. Som exempel, om du tyckte att det var lite jobbigt, så kryssar du i rutan framför "lite jobbigt". Titta på exemplet nedan.

Fråga: Hur ofta har du under de senaste 7 dygnen haft löst bajs?

aldrig ibland ofta mycket ofta

Hur jobbigt tyckte du att det var?

Inte alls jobbigt lite jobbigt jobbigt mycket jobbigt

Om du vill ändra ditt svar, kryssa så över "fel" svar och ringa in "rätt" svar

Så här: ~~aldrig~~ ibland **ofta** mycket ofta

Ringa in det svar som passar bäst in på dig. När du fyllt i en hel fråga så kan du fortsätta till nästa fråga

				CF 8-11år
1. Hur ofta under de senaste 7 dygnen åt du något med flit för att göra ditt bajs löst?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
2. Hur ofta under de senaste 7 dygnen åt du något med flit för att göra ditt bajs fast?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
3. Hur ofta under de senaste 7 dygnen lät du bli att äta viss mat för att du inte skulle få löst bajs	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
4. Hur ofta under de senaste 7 dygnen lät du bli att äta viss mat för att du inte skulle få hårt bajs?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
5. Hur ofta under de senaste 7 dygnen kom det lite kiss innan du hann till toaletten?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
6. Hur ofta under de senaste 7 dygnen kom det kiss i byxorna när du lekte, cyklade, eller sprang?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				

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	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
7. Hur ofta under de senaste 7 dyggen, kom det kiss i byxorna när du var rädd, arg eller glad?				
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
8. Hur ofta under de senaste 7 dyggen kom det kiss i byxorna när du hostade eller nös?				
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
9. Hur ofta under de senaste 7 dyggen var det mycket viktigt för dig att vara nära en toalett?				
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
10. Hur ofta under de senaste 7 dyggen behövde du gå ifrån klassrummet för att du var tvungen att gå på toaletten?				
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
11. Hur ofta, under de senaste 7 dyggen kände du att du blev mer retad än andra barn på grund av dina besvär?				
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
12. Hur ofta, under de senaste 7 dyggen kände du att du var mindre söt än andra barn på grund av dina besvär?				
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				

13. Hur ofta, under de senaste 7 dygnen var du inte nöjd med din kropp?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				

14. Hur ofta under de senaste 7 dygnen, har du känt dig generad över dina besvär?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				

15. Hur ofta, under de senaste 7 dygnen, har du tänkt att du är annorlunda jämfört med andra barn på grund av dina besvär?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				

16. Hur ofta under de senaste 7 dygnen tänkte du att andra tyckte att du var mindre söt/trevlig på grund av dina besvär?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				

## 17. Har du stomi?

**Nej** ⇒ Var vänlig och svara på fråga 18 till 46.  
(Du kan hoppa över fråga 47 till 58)

**Ja..⇒** Var vänlig och svara på fråga 47 till 58.  
(Du kan hoppa över fråga 18 till 46)

Följande frågor handlar om hur ofta du har bajsat under de senaste 7 dygnen.

Var snäll och ringa in det svar som du tycker passar bäst in på dig.



18. Av hur många dagar av de senaste 7 dygnen kunde du bajsas av dig själv?  
(alltså utan att använda något medel för att hjälpa dig bajsas, men du ska inte räkna när du har bromsspår i underkläderna)

A. 0 dagar, inte en enda gång under de senaste 7 dygnen  
B. 1 dag  
C. 2 dagar  
D. 3 dagar  
E. 4 dagar  
F. 5 dagar  
G. 6 dagar  
H. 7 dagar

19. Av hur många dagar av de senaste 7 dygnen bajsade du mer än 4 gånger på en dag?

A. 0 dagar, inte en enda gång under de senaste 7 dygnen  
B. 1 dag  
C. 2 dagar  
D. 3 dagar  
E. 4 dagar  
F. 5 dagar  
G. 6 dagar  
H. 7 dagar

20. Av hur många dagar av de senaste 7 dygnen kände du dig förstoppad och kunde inte få ut allt bajs?

A. 0 dagar, inte en enda gång under de senaste 7 dygnen  
B. 1 dag  
C. 2 dagar  
D. 3 dagar  
E. 4 dagar  
F. 5 dagar  
G. 6 dagar  
H. 7 dagar

21. Vad använde du under de senaste 7 dagarna för att kunna bajsas?  
(Här får du ringa in mer än ett svar)

A. Inget  
B. Tarmsköljning  
C. Minilaxering (Resulax, Klyx)  
D. Medicin i munnen (t.ex. Movicol Importal etc.)  
E. Annat \_\_\_\_\_  
F. Jag vet inte

22. Om du under de senaste 7 dagarna har använt något medel för att kunna bajsas så var det  
(Här får du ringa in mer än ett svar)

- A. För att du var förstoppad
- B. För att du inte ville läcka bajs
- C. För att du bajsade för lite
- D. För att ditt bajs var för hårt eller för löst
- E. För att \_\_\_\_\_
- \_\_\_\_\_
- F. Inte tillämplig, för jag använde inget
- G. Jag vet inte

23. Av hur många dagar under de senaste 7 dyggen har du bajsat väldigt mycket på en gång?

- A. 0 dagar, inte en endaste gång under de senaste 7 dyggen
- B. 1 dag
- C. 2 dagar
- D. 3 dagar
- E. 4 dagar
- F. 5 dagar
- G. 6 dagar
- H. 7 dagar
- I. Inte tillämplig, för jag bajsar aldrig mycket vid ett tillfälle

Följande frågor handlar om hur ofta vissa saker har förekommit under de senaste 7 dyggen

Var snäll och ringa in det svar som passar bäst in på dig.

24. Hur ofta, under de senaste 7 dyggen har ditt bajs varit löst?

Aldrig    Ibland    ofta    Mycket ofta  
↓        ↓        ↓        ↓

Hur jobbigt tyckte du att det var?

☐ Inte alls jobbigt   ☐ Lite jobbigt   ☐ Jobbigt   ☐ Mycket jobbigt

25. Hur ofta under de senaste 7 dyggen var ditt bajs löst mer än 4 gånger på en dag?

Aldrig    Ibland    Ofta    Mycket ofta  
↓        ↓        ↓        ↓

Hur jobbigt tyckte du att det var?

☐ Inte alls jobbigt   ☐ Lite jobbigt   ☐ Jobbigt   ☐ Mycket jobbigt

26.	Hur ofta under de senaste 7 dyggen var ditt bajs hårt?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?					
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt					

27.	Hur ofta under de senaste 7 dyggen kändes det som det var fullt i magen?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?					
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt					

28.	Hur ofta under de senaste 7 dyggen kände du dig inte bajsning fast det fanns bajs i din mage?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?					
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt					

29.	Hur ofta under de senaste 7 dyggen gick du på toaletten för att bajs utan att känna att bajset var på väg?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?					
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt					

30.	Hur ofta under de senaste 7 dyggen var det svårt att få ut bajset?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?					
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt					

31.	Hur ofta under de senaste 7 dyggen har du pruttat utan att veta om det innan (att prutten var på väg)?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?					
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt					

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32. Hur ofta under de senaste 7 dyggen pruttade du?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
33. Hur ofta under de senaste 7 dyggen var det svårt att prutta?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
34. Hur ofta under de senaste 7 dyggen lät det mycket från din mage (knorrat/bullrat)?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte jobbigt alls <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
35. Hur ofta under de senaste 7 dyggen hade du ont i magen?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
36. Hur ofta under de senaste 7 dyggen läckte det bajs innan du kom fram till toaletten?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
37. Hur ofta under de senaste 7 dyggen, fick du bromsspår i byxorna under dagen?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				

38. Hur ofta under de senaste 7 dyggen har  
fick du bromsspår i byxorna under natten?

Aldrig    Ibland    Ofta    Mycket ofta  
↓           ↓           ↓           ↓

Hur jobbigt tyckte du att det var?

☐ Inte alls jobbigt   ☐ Lite jobbigt   ☐ Jobbigt   ☐ Mycket jobbigt

39. Hur ofta under de senaste 7 dyggen  
bajsade du på dig lite grann under natten?

Aldrig    Ibland    Ofta    Mycket ofta  
↓           ↓           ↓           ↓

Hur jobbigt tyckte du att det var?

☐ Inte alls jobbigt   ☐ Lite jobbigt   ☐ Jobbigt   ☐ Mycket jobbigt

40. Hur ofta under de senaste 7 dyggen  
bajsade du på dig lite grann när du lekte,  
cyklade eller sprang?

Aldrig    Ibland    Ofta    Mycket ofta  
↓           ↓           ↓           ↓

Hur jobbigt tyckte du att det var?

☐ Inte alls jobbigt   ☐ Lite jobbigt   ☐ Jobbigt   ☐ Mycket jobbigt

41. Hur ofta under de senaste 7 dyggen  
bajsade du på dig lite grann när du var rädd,  
arg eller glad?

Aldrig    Ibland    Ofta    Mycket ofta  
↓           ↓           ↓           ↓

Hur jobbigt tyckte du att det var?

☐ Inte alls jobbigt   ☐ Lite jobbigt   ☐ Jobbigt   ☐ Mycket jobbigt

42. Hur ofta under de senaste 7 dyggen  
bajsade du på dig lite grann när du hostade  
eller nös?

Aldrig    Ibland    Ofta    Mycket ofta  
↓           ↓           ↓           ↓

Hur jobbigt tyckte du att det var?

☐ Inte alls jobbigt   ☐ Lite jobbigt   ☐ Jobbigt   ☐ Mycket jobbigt

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43. Hur ofta under de senaste 7 dyggen har du varit rädd att dina kompisar skulle känna att det luktade bajs om dig?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
44. Hur ofta under de senaste 7 dagarna stannade du hemma från skolan för att du var rädd att du skulle bajs på dig lite grann?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
45. Hur ofta under de senaste 7 dagarna ville du gå ut och leka eller leka hemma hos en kompis, men stannade hemma för att du var rädd för att smutsa ned underkläderna?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				
46. Hur ofta under de senaste 7 dagarna ville du idrotta men stannade hemma för att du var rädd för att bajs på dig?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?				
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt				

Följande frågor (frågorna 47 till 58) handlar om barn med stomi.

Försök att svara på alla frågor och hoppa inte över någon fråga

Ringa in det svar som passar bäst in på din situation.

47. Hur ofta under de senaste 4 veckorna har du haft något av följande problem med din stomi?  
(Du får ringa in mer än ett svar)

A. Prolaps  
B. Förträngning (trångt) i stomin  
C. Hud irritation  
D. Annat \_\_\_\_\_  
E. Nej, jag har inte haft problem med min stomi

48. Tycker du att din stomi är bra placerad/ sitter på ett bra ställe?

A. Ja  
B. Nej

Om du inte tycker att din stomi är bra placerad, skriv varför du inte tycker det

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49. Vad har du använt under de senaste 7 dagarna för att göra så att det ska komma mindre eller mer bajs ut ur stomin?  
(Du får ringa in mer än ett svar)

A. Ingenting  
B. Sköljning  
C. Klyx  
D. Medicin i munnen  
E. Annat \_\_\_\_\_  
F. Jag vet inte

50. Om du har använt något för att det skall komma mer eller mindre bajs ur din stomi, var det isåfall för att: ( Du får ringa in mer än ett svar)

A. Det kom för mycket bajs  
B. Det kom för lite bajs  
C. Du ville inte att det skulle komma bajs  
D. Annat (var snäll och skriv vad) \_\_\_\_\_

E. Jag använde ingenting för att det skulle komma mer eller mindre bajs ur min stomi

Följande frågor handlar om **hur ofta** vissa saker har hänt under de senaste 7 dagarna.  
 Var snäll och ringa in det svar som passar bäst in.

51.	Hur ofta under de senaste 7 dagarna var ditt bajs löst?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?					
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt					
52.	Hur ofta under de senaste 7 dagarna läckte din stomipåse under dagen?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?					
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt					
53.	Hur ofta under de senaste 7 dagarna läckte din stomipåse på natten?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?					
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt					
54.	Hur ofta, under de senaste 7 dagarna var du orolig för att dina vänner skulle känna lukten av ditt bajs?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?					
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt					
55.	Hur ofta, under de senaste 7 dagarna var du orolig för att dina vänner skulle se din stomi?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?					
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt					
56.	Hur ofta, under de senaste 7 dagarna var du orolig för att dina vänner skulle höra att det lät från din stomi?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
Hur jobbigt tyckte du att det var?					
<input type="checkbox"/> Inte alls jobbigt <input type="checkbox"/> Lite jobbigt <input type="checkbox"/> Jobbigt <input type="checkbox"/> Mycket jobbigt					
57.		Aldrig	Ibland	Ofta	Mycket ofta



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Hur ofta under de senaste 7 dagarna var du orolig för att din stomipåse skulle börja läcka?	↓	↓	↓

Hur jobbigt tyckte du att det var?

☐ Inte alls jobbigt ☐ Lite jobbigt ☐ Jobbigt ☐ Mycket jobbigt

58.	Hur ofta under de senaste 7 dagarna, har du haft problem när du sköter om din stomi/tar hand om din stomi?	Aldrig	Ibland ↓	Ofta ↓	Mycket ofta ↓
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Hur jobbigt tyckte du att det var?

☐ Inte alls jobbigt ☐ Lite jobbigt ☐ Jobbigt ☐ Mycket jobbigt

Var det något du tyckte var svårt eller konstigt när du fyllde i enkäten?

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Är det något annat du tänker på?

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English translations A/Prof AGCatto-Smith & M Trajanovska  
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